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EFFECTIVE MANAGEMENT OF PEMPHIGUS VULGARIS: A CASE REPORT

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Abstract

Pemphigus vulgaris is a unique chronic condition of bullous autoimmune dermatosis whose development and diagnosis is mostly unidentifiable. It is classified as a specific hypersensitivity type II reaction that initiates the antibodies against the desmosomes, which are the maculae adherentes of the skin also known as the adhering spots in common term. These adhering spots keep the certain layers and compartments of the skin intact. When the desmosomes are invaded by the antibodies, the intact layers of the skin separate out and the clinical picture resembles a blister. With the passage of time the condition becomes worse and without treatment and monitoring the blister type lesions increase in size and surface area of the body similar to the condition of severe burn. The prime focus in the treatment of pemphigus vulgaris is to control the infirmity, prevent relapses, avoid adverse events and complications associated with the broad use of the steroids and immunosuppressive agents. Systemic corticosteroids evidenced as the golden standard in the treatment regimens of the pemphigus vulgaris. Azathioprine and mycophenolate mofetil are considered as the choice of drug in the treatment of the pemphigus vulgaris as a steroid sparing agent. Rituximab is believed to be immensely efficacious in unmanageable and refracted types of pemphigus vulgaris when other remedies fail to control the disease. One of the most noticeable complications of the pemphigus vulgaris is the compromised quality of life due to the blister forming all over the body which aggravates the soreness and pain on the affected area leading to ulceration and formation of erosive lesions. If there is a recurrence of the disease then the steroids doses will be checked properly and if needed then the doses could also be increased according to the weight in safe limit thus augmenting the immunosuppressants in the body and find an alternate safe drug in concomitant therapy

Keywords: *Corticosteroids, Erosions, Immunosuppressant, Lesions, Pemphigus vulgaris, Treatment*

INTRODUCTION

The term “Pemphigus” is derived from the Greek word pemphix means bubble or blister (1) and the term “Vulgaris” is derived from the Latin word means common (2) or something that is derived from the masses of common people. Pemphigus is probably a life threatening disease that causes erosion and fluid filled vesicles on the skin and mucous membrane (3). Pemphigus Vulgaris is a chronic , infrequent, intraepidermal bullous disease with potentially lethal consequences and initially named by Wickman in 1791. Pemphigus is an unusual disease with the incidence rate ranging from 0.5 - 3.5 cases per 100,000 annually. The mortality rate of the cases with pemphigus vulgaris is approximately 5-15% of the total population (4). Above all pemphigus vulgaris accounts for approximately 70% of all the types of the pemphigus cases.

Pemphigus Vulgaris is considered as an organ specific autoimmune disease indicated by the assembly of the autoantibodies directed against the desmosomal proteins that advances to acantholysis and thus forming the epidermal bullae which presents an image of blister on the body (5,6). The prognosis of the pemphigus vulgaris had revolutionized the world by introducing corticosteroids which significantly declined the mortality rate from 75-80% to only 30%, which was a remarkable invention. Even with the disease still associated with a mortality rate of about 6% in spite of the use of the various adjuvant treatments and remedies (7, 8).

The disease is characterized by the flaccid blisters that rupture easily, leaving painful erosion with severe burning sensation (9). Baun et al. observed that the clinical representation of the pemphigus vulgaris is 41.7% of mucosal lesions, 37.4% of simultaneous mucosal and 20.4% of the cutaneous lesions appeared on

the body (10). It is properly diagnosed on the basis of examination of a biopsy specimen with hematoxylin-eosin staining and direct immunofluorescence for the accurate and better results (11).

Before the initiation of the systemic corticosteroids the mortality cases with pemphigus vulgaris ranged between 60-90% (12) but now a big gratitude to the usage of the corticosteroid sparing agents, the mortality rate reduced and stands around 10% (13).

The neglected untreated case of pemphigus vulgaris is quite fatal and delayed in the management leads to the involvement of the other vital organs. That is the main reason to identify the lesions and it is the prior responsibility of the clinician to make differential diagnosis as soon as possible for early treatment without wasting any time which is of paramount prognostic importance (14).

CASE STUDY

A 40 years old married male, farmer by occupation, resident of Jatoi complained of multiple vesicles and blisters all over his body. Patient was in his usual state of health 5 months back from now when he developed fluid filled vesicles and blisters. These are flaccid, containing purulent fluid starting from the back and progressing towards the face, scalp, trunk, genitalia, and limbs within 10 days. It can easily rupture within 1-2 hours spontaneously or by friction leaving behind painful erosions associated with itching and burning sensation. These are aggravated by sun exposure and retrieved by taking antihistamines temporarily. A patient also developed multiple erosions in the oral cavity associated with difficulty in eating, drinking and swallowing as well. Moreover, there is no history of fever, cough, short of breath and burning micturition. Patient had already taken treatment from the quacks i.e. hakims which aggravated the condition and made his condition fatal.

Now, there are many intact blisters on his back and trunk, multiple erosions all over the body and developed yellowish crust predominantly on forehead, scalp and back along with multiple erosions in oral cavity and genitalia (Fig. 1a & b). Palms and soles are spared, nails are pallor but normal. SOAP analysis was followed on that patient regularly from the date of admission. Patient was administered with the following pharmacological plan with effective and speedy prognosis (Table I). Along with the pharmacological treatment, nursing care was the most important factor in the effective and positive management of the pemphigus vulgaris.

Table I. Pharmacological plan for the patient

Name of drugs	Dosage	Name of drugs	Dosage
Calamox (Injection)	1.2 gm	Qalsan D (Tablet)	1 (OD)
Azithromycin (Injection)	500 mg	Sangobion (Tablet)	1 (OD)
D- Tres (Injection)	2 lac	Omega (Capsule)	40 mg
Decadron (Injection)	2 cc	Polyfax skin Ointment	BD
Rituximab (Injection)	500 mg	Liquid Paraffin + Plain Vaseline	
Polyfax eye ointment	on both eyes	Enziclor mouthwash + Nilstat oral drops + Glycerin	

After 18 days of the stay in hospital with the management the patient moved toward normal physiology and the eruption of the new blisters and lesions also stopped. The crust also started to form on some of the erosion. The eruption of new blisters stopped after the administration of the Rituximab 500 mg doses with close monitoring. This drug is considered as the magical drug in the treatment of the aggravated case of the pemphigus vulgaris. Fig. 1 (c) shows the effectiveness of the rituximab doses.

DISCUSSION

Pemphigus vulgaris is a fatal and chronic condition if left untreated. It badly deteriorates the quality of life due to the erosive skin texture, burning sensation and perception of the pain of scale 7-8 out of 10 as verbalized by the patients. The nutritional value of the patient is also badly compromised due to the ulceration of the oral mucosa, so the patient is unable to eat and drink as recommended and thus become malnourished.

In the management of the pemphigus vulgaris, the action of the corticosteroids are evidenced as a golden standard for the treatment and thus declining the mortality rate globally. The initial dose of systemic

corticosteroids is experiential and is generally based on clinical proficiency and intensity of the disease. It commonly ranges from 0.5 mg/kg/day to 2 mg/kg/day in a safe limit as recommended (15). In spite of the therapeutic use, the increased doses of steroids and its advantages over dosages of 1 mg/kg/ day is still vague and further study on it is a demand of the present time (16).

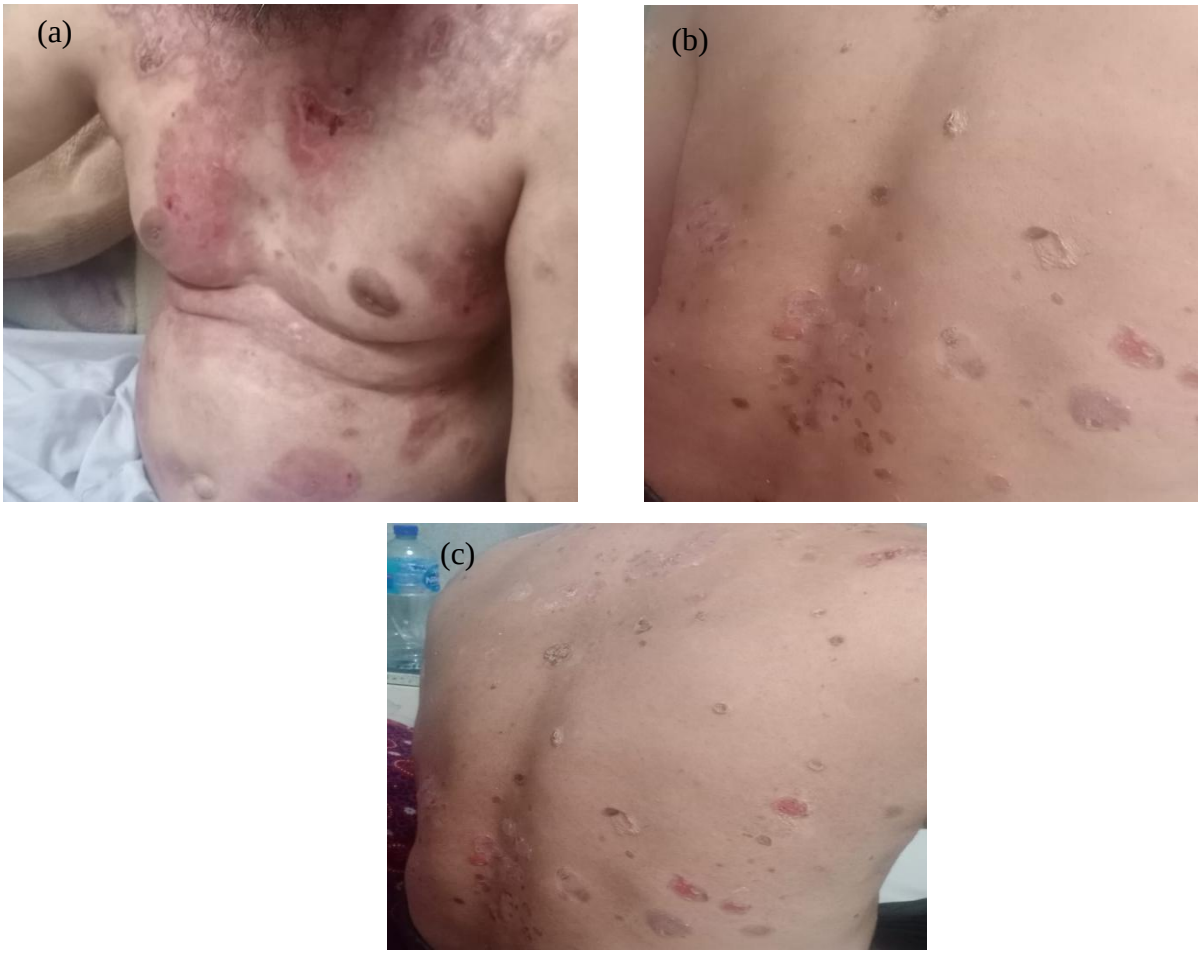

Fig. 1. (a) shows the involvement of the trunk, (b) shows the involvement of the back and (c) shows the formation of the yellowish crust on the blisters

Immunosuppressive medications are the potent steroid sparing agents and it needs keen consideration before the expected time in the management of the disease. Epidermal growth elements aid to speed up the recuperation of the ulcerative lesions on the body (17). Corticosteroid sparing agents i.e. methotrexate, azathioprine, mycophenolate mofetil (MM) and rituximab are the potent combination and augmented the efficacy of the systemic corticosteroids thus decreases the need of these medications and preventing from the inauspicious effects (18).

Multiple authorities nowadays rely on the injectable doses of rituximab as the first and second line agents with the evidence from the study conducted in 2017. As a first line agent rituximab showed better and acceptable results in the management of the pemphigus vulgaris (19). Injectable rituximab was ratified by the US Food and Drug Administration (FDA) in the treatment plan of the pemphigus vulgaris in June 2018. On the basis of the comparative randomised trial performed between the efficacy of the rituximab along with oral corticosteroids and alone corticosteroids as a first drug of choice in the cases of pemphigus vulgaris with moderate to severe range. The result showed the better therapeutic result of the injectable rituximab doses, through this study it was approved by the competent authority of the FDA to the therapeutic use of the rituximab injectable as a drug of choice (20).

The antitumor necrosis factor drugs i.e. sulfasalazine and pentoxifylline have been revealed as one of the efficacious concomitant management thus decreases the serum level of the tumor necrotizing factor which proves helpful in the speedy recovery of the pemphigus vulgaris (21). Methotrexate also showed therapeutic effects.

In the severe cases of the pemphigus vulgaris, intravenous pulse therapy was also highly recommended for some duration with methylprednisolone, cyclophosphamide or gamma globulin to lessen the ulcerative effects of the blisters formed all over the body (22). Cyclophosphamide is also considered as

the effective drug in the refractory phase of the pemphigus vulgaris. Above all the role of the causative agent is still a mystery and being investigated (23).

Topical remedy for cutaneous aid in pemphigus vulgaris may include topical steroids and emollients. It proves really helpful along with the mouthwash and oral steroids for the oral ulceration.

CONCLUSION

Pemphigus vulgaris is a unique cause of the chronic erosive ulceration of the mucosal membrane in about 80% of the cases. There is a changing trend of the severity and natural history of the disease among the different cases, the effects of the disease are not same thus clinical presentation of the different cases are vacillating. In the efficient management of the pemphigus vulgaris, it is very essential to make the differential diagnosis in time so that prompt treatment could be initiated which saves people from the aggravated phase of the pemphigus vulgaris and thus decreases the mortality and morbidity rate worldwide. Untreated cases of pemphigus vulgaris are more susceptible to the infection and deranged fluid and electrolyte levels. Through the studies it is revealed that this disease demands a high coping spirit. The body image in this disease is highly compromised and most of the people cannot cope with it and die in the initial years but those having high coping spirit and strong supportive background, they will survive. People who survived more than 5 years have a good prognosis. Supportive treatment along with the pharmacological interventions and nursing interventions speed up the rate of the recovery as this disease deteriorates the body image and it requires too much patience and cooperation of the patient as well as their families to cope with the challenges faced during the disease process.

Consent for Publication:

Written informed consent was taken from the patient for study and publication as well. None of his personal information will be disclosed in the final publication.

Conflict of interest:

The author reports no conflict of interest.

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